



DPT

DEVELOPMENTAL PEDIATRICS TODAY



August 2020

Monthly e-Newsletter of IAP Chapter of Neurodevelopmental Pediatrics

IAP CHAPTER OF NEURO DEVELOPMENTAL PEDIATRICS

Chairperson : Dr Shabina Ahmed
Hon'Secretary : Wg Cdr (Dr) KS Multani
Past Chairperson : Dr Jeelon C. Unni
Past secretary : Dr Leena Sreevastava
Joint secretary : Dr Zafar Meenai
Dr Shambhavi Seth
Dr Arun Prasad
Treasurer : Dr Muhamed Ismail P.M.

ADVISORS

Dr MKC Nair
Dr SS Kamath
Dr Abraham K. Paul
Dr Pratibha Singhi
Dr Nandini Mundkur

National coordinator of the TOT program

Dr Samir Dalwai

National coordinator of the Fellowship program

Dr Chhaya Prasad

EDITORIAL BOARD

Chief Editor : Dr Lata Bhat
Editorial Board : Dr Jyoti Bhatia
Dr Anjan Bhattacharya
Dr Leena Deshpande
Dr Anju Agarwal
Dr Arun Prasad
Dr Sivaprakasam

WEBSITE COMMITTEE

Dr Zafar Meenai, Dr Somasundaram, Dr Lata Bhat
Dr P Sudhakar, Dr Mahesh Mohonto

STATE CO-ORDINATORS

Armed Forces : Wng Cdr Kawaljit Singh Multani
Andhra Pradesh : Dr Hanumantha Rao
Assam : Dr Sujit Kumar Chaudhary
Bihar : Dr Anil Kumar Tiwari
Dadra Nagar : Dr Sunil Datt P Daru
Haveli Silvassa
Delhi /NCR : Dr Lata Bhat
Goa : Dr Aparna Shirodkar
Gujarat : Dr Swati Vinchurkar
Haryana : Dr Harsh Bhayana
HP : Dr Ashwini Sood
Karnataka : Dr M Mahadeviah
Kerala : Dr Jacob Roy
Madhya Pradesh : Dr Jyotsna Shrivastava
Maharashtra : Dr Leena Srivastava
Orissa : Dr Mahesh Mohanta
Rajasthan : Dr S. Sitaraman
Tamil Nadu : Dr A. Somasundaram
Telangana : Dr Namratha Rao
Uttar Pradesh : Dr Alka Agarwal

Inside

Editorial.....	2
Chairpersons Message.....	3
Snippets from the Secretary	4
Breastfeeding - Origins, Present & Future	5
Developmental Coordination Disorder.....	7
Case Report	9
Journal Scan.....	13
Quiz.....	16
Month in pics.....	18



Editorial

Dear Friends and respected Seniors,



Hope you all are fine. There is an increase in Covid cases in India and our country has leaped to number 2 position in terms of total cases next only to USA. Yet life is slowly creeping back to near normal with gradual unlock happening. With adequate preventive measures, Child development centers are giving therapies, but the online therapies are here to stay.

We feel proud that our chapter is continuing to contribute to dIAP webinars.

August is awareness month for – ‘Children’s eye health and safety’, ‘Child vision and learning’, ‘Spinal muscular atrophy awareness month’ and ‘Don’t be a bully awareness month’. All these are important to spread awareness because they have immense contribution to child development. However, I would like to stress upon awareness regarding bullying, because in India most schools don’t have a system to prevent bullying and there is lack of awareness amongst teachers as well as parents. All of us should keep a high index of suspicion about bullying and screen for the same in our clinics.

We are giving quiz questions and I would request you all to send your answers by 25th September 2020. We are publishing case reports and will continue to do so. Again, a humble request to all of you to send interesting cases to us and also send your answers for the quiz.

There has been an increase in the number of Covid cases, so please don’t forget the three main safety precautions against Covid19 - mask, social distancing and hand sanitization. Stay safe, stay healthy.

Dr. Lata Bhat

Chief Editor



Chairperson's Message

Dear Readers,

As the calendar is rolling by, every month of the year has a message, a theme for celebration of its importance and reminder of sustaining these activities for posterity. All for the good health of the inhabitants and a healthy planet



The members of Neurodevelopmental Chapter are putting in their efforts to bring to readers new information and excitement in the understanding of the neurodevelopmental problems and promotion of care.

This month our focus is on World Breast Feeding Week and we are specifically concerned because of the link of breastmilk and neurodevelopment. It has short-term and long-term benefits which are of interest to clinicians and policy makers. The advent of "Proton Magnetic Resonance Spectroscopy" has helped the researchers to peep inside the brain of infants and have found signature biochemicals like inositol, creatinine, choline which are associated with improved intelligence and working memory in breastfed toddlers.

Breastfeeding is a natural process but it needs support both to start and to sustain, and it is our duty to provide access to a woman, skilled breastfeeding counselling. This is in line with this year's WHO theme, "Support breastfeeding for a healthier planet".

This issue gives you important insight related to the above topics of importance of this month.

Happy reading.

Long Live IAP !

Dr. Shabina Ahmed MD, FIAP

National Chairperson

Neurodevelopmental Pediatrics Chapter of IAP



Snippets from the Secretary

“A newborn baby has only three demands - warmth in the arms of its mother, food from her breasts and security in the knowledge of her presence. Breastfeeding satisfies all three.”

- Grantly Dick-Read



Respected seniors and dear friends,

Greetings from the IAP chapter of Neurodevelopmental pediatrics.

Hope this issue of newsletter finds you all in good health. We are now in the sixth month of COVID-19 pandemic in India and the number of cases in the country are rapidly increasing with India reaching the number two spot in the worst affected countries in the world in terms of number of cases. Luckily, the number of fatalities in the country is low compared to the world data. The role of all personnel involved in the fight against the pandemic is praiseworthy and has highlighted the need for improvement of basic medical infrastructure in the country. Hope the lessons from the pandemic are used for improvements in the health infrastructure and facilities at the primary level by the government.

The month started with the world breastfeeding week (01-07 Aug) and the theme for this year is ‘Support breastfeeding for a healthier planet’. We have a few interesting articles on breastfeeding in this issue and an article on the past and present of breastfeeding. Breastfeeding, which is considered as the first vaccine for the child, is also the best source of nutrition for the newborn. We have been slowly moving away from breast-feeding due to diet fads and other misconceptions which is evident from low rates of exclusive breastfeeding all over the world. We all should wholeheartedly support this noble cause for a safe and bright future for our children and the coming generations.

The neurodevelopment chapter runs a one year fellowship in ‘Developmental and Behavioral Pediatrics’ (under the aegis of central IAP) at 12 centre all over the country for the last few years and this year, we have added three new centers to the list of existing centers and now, we have also started a nursing fellowship in ‘Developmental and Behavioral Pediatrics’. The chapter has been active on the academic front in the activities of dIAP and we thank the IAP President and Secretary in their efforts of keeping all of us connected and academically oriented in these difficult times.

As we enter into the fourth phase of Unlockdown, we wish you all a safe month ahead.

“We cannot always build the future for our youth, but we can build our youth for the future.”

- Franklin D. Roosevelt.

Jai Hind!

Wg Cdr (Dr) KS Multani

National Secretary

IAP Chapter of Neurodevelopmental Paediatrics



Breastfeeding - Origins, Present & Future

Wg Cdr (Dr) KS Multani

Mammals as a group in the animal kingdom are characterized by their unique ability to give birth and nurse their young ones. Mammals include a wide variety of animals from rats to dogs to elephants to seals and dolphins as well as humans. Lactation is a primal behaviour and has its origins hundreds of millions of years in the past(1). Breastfeeding



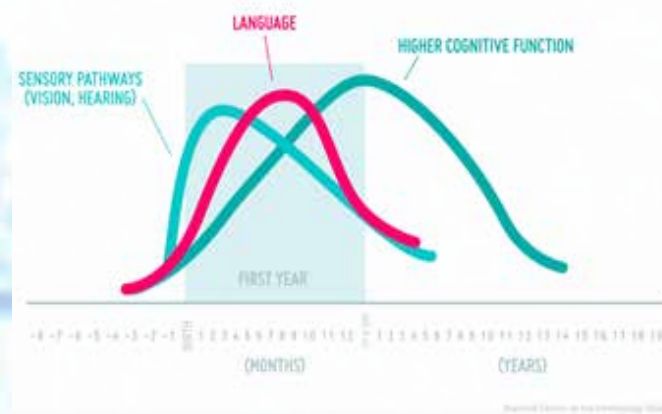
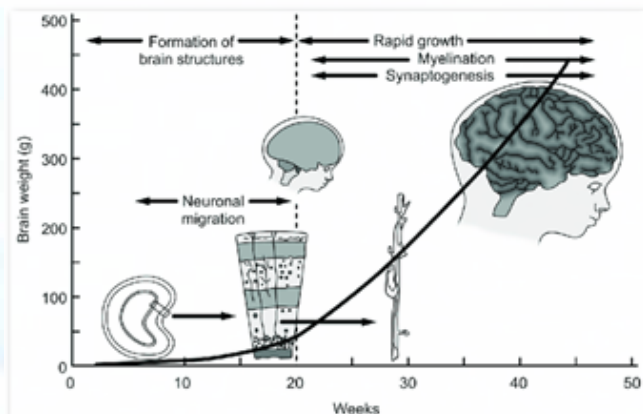
is an intimate, emotional experience and has beneficial effects for both - the mother and the child. Breastmilk is the best food for the baby best suited according to the animals needs as the milk composition of different animals is determined by the species needs. In case of humans, the milk contains special components having role in brain development in addition to other nutrient and protective factors(see journal scan for some selected articles on this subject).

Human beings are distinct from other animals due to their large brain size and higher cognitive thinking but this same reason also makes the newborn at a disadvantage during early life. Though the brain of a newborn is structurally complete on a macro level at the time of birth, it is still undergoing lot of structural changes at microscopic level (neuronal migration, synapse formations, myelination and synapse sculpting) which are essential for the development of higher brain functions later in life like language

and cognitive thinking (see image) and thus, has an impact on the person's performance later in life. Breastfeeding has a major effect on development of language and higher cognitive function in life as it has special components which ensure and promote the ongoing brain development activities which has been demonstrated in many studies that have shown

that breastfed children have higher IQ compared to non-breastfed children.

Sadly, over time humans have moved away from exclusive breastfeeding of their young ones which is evident from low rates of exclusive breastfeeding in both developed and developing nations. Hence, a need was felt to protect and support breastfeeding and a declaration towards this was made by World Health Organization (WHO) and United Nations Children's Fund (UNICEF) in 1990 - 'the Innocenti Declaration'. World alliance for breastfeeding action (WABA), which was formed in 1991, is a global alliance network of individuals and organizations dedicated to the protection, promotion and support of breastfeeding worldwide. WABA collaborates with the WHO and UNICEF to make sure that aid is given to the right people, in the right communities. World breastfeeding week (WBW) is celebrated every year in the first week of August world over and it was first celebrated



in 1992. WBW aims to highlight the huge benefits that breastfeeding can bring to the health and welfare of babies and benefits to maternal health, focusing on good nutrition, poverty reduction, and food security. World breastfeeding week has the dual goal of improving the health of babies and promoting, protecting, and supporting the rights of women to breastfeed anywhere and at any time. IMS Act of 1992 (Infant milk substitutes, feeding bottles and infant foods) was passed by Indian government in 1992 to protect breastfeeding by controlling the marketing and promotional activities of baby food manufacturers and also ensuring proper use of breast milk substitutes. An amendment was made in the act in 2003 to increase the scope of the act. The act prohibits the companies to use pictures of women/infant on the baby food labels and instructs them to have a label in english and local language that 'Breastfeeding is the BEST' on their products and gives clear guidelines for educational materials and advertisements dealing with pre/post natal care and infant feeding. The act also provides for monetary fines and imprisonment penalties for violators.

Maternal education status, type of delivery, low birth weight and NICU stay are some factors

that have been found to be linked with exclusive breastfeeding in a recent study (see journal scan). This year the theme chosen for WBW is 'Support breastfeeding for a healthier planet'. WBW 2020's goals have been aligned with United Nations Sustainable Development Goals (SDGs) and WABA is applying the 'warm chain' approach to empower women from all sectors of the world to unite and work towards the singular goal of protecting the environment. WABA talks about how **"Breastfeeding is a prime example of the deep connections between human health and nature's ecosystem. Breast milk is a natural, and renewable source of nourishment for children that does not adversely impact the environment and is completely green as it produces no waste, and its production and delivery causes no pollution."** Through the WBW 2020 campaign, WABA also means to highlight the negative effects that artificial feeding has on the environment and how it serves as a drain on natural resources.

Further reading :

1. The origin and evolution of lactation. Anthony VC, RM Akers. J Biol. 2009; 8(4): 37.



Developmental Coordination Disorder

DR. ASHISH SAHANI

Developmental coordination disorder (DCD) is characterized by problems with motor coordination that interfere with academic performance and social integration in otherwise healthy children. It typically presents in the early school years and persists into adolescence or adulthood.

ETIOLOGY

Etiology is unknown. Factors related to the task (complexity, pace), environment (visual cues, opportunity for practice), and individual child appear to contribute.

Factors related to the child include motivation, perseverance, difficulty in executing motor tasks, problems with proprioception and sensory integration, genetic factors and atypical brain development. Atypical brain development is supported by neuroimaging studies. In DCD there is less activation of brain regions responsible for motor automaticity and differences in the parieto-frontal connections (which integrates visuo-spatial information) as decoded by MRI studies.

EPIDEMIOLOGY

PREVALANCE: DCD is two to seven times more common in boys than girls. The prevalence is not affected by socio-economic status or educational level. The prevalence varies from 1.7 percent to 6 percent.

RISK FACTORS: DCD is strongly related to history of preterm and low birth weight. According to one study, administration of caffeine therapy for apnea of prematurity may reduce risk of DCD in

preterm infants.

ASSOCIATED CONDITIONS: DCD is associated with other neurodevelopmental conditions like ADHD, ASD, specific language impairment, dyslexia and other behavioural problems.

CLINICAL FEATURES

CHILDREN: The diagnosis is made between 6 to 12 years. These children typically have delays in achieving motor skills throughout life, but they become more apparent when it interferes with socio-adaptive development. Parents may complain that children have difficulty tying shoe laces or brushing teeth, playing catch or riding a bicycle. There are delays in self-care skills. In school, there is crude pencil grasp, poor handwriting, and inability to cut paper in a straight line. Child may bump into walls or classmates and may cause a lot of social problems for the child.

ADOLESCENTS

Motor difficulties persist into adolescence in 50% of children. They face difficulty in hand skills, agility, balance, driving and playing sports. Co-existing non-motor problems include difficulty with attention, and executive function, anxiety, obesity and poor general health.

DSM-5 criteria for diagnosis of DCD:

1. The achievement and performance of coordinated motor skills is substantially below that expected given the child's chronological age and opportunity for skill learning and use.



2. The poor performance significantly and persistently interferes with activities of daily living appropriate to chronologic age and impacts academic/school productivity, pre-vocational and vocational activities, leisure and play.
3. The symptoms began in early development period.
4. The impairments in motor skills deficits are not better explained by intellectual disability or visual impairment and cannot be attributed to another neurologic or neuromuscular condition affecting movement.

Although age >5 years is not a diagnostic criterion, the diagnosis of DCD is rarely made in children less than 5 years unless they have severe impairment.

Weakness of limbs, ataxia, pronounced hypotonia or hypertonia and increased or decreased DTRs particularly when asymmetric are consistent with DCD.

Lab and imaging studies are not usually necessary in a child of suspected DCD but may be warranted in children with acute or on progressive changes in gross motor skills or abnormal findings on neurological examination. LDH and creatine kinase may be warranted in children with poor muscle mass.

DIFFERENTIAL DIAGNOSIS

Non progressive incoordination:

1. Intellectual disability
2. ADHD
3. ASD
4. Vertigo
5. acquired brain injury

6. neonatal encephalopathy
7. orthopedic and rheumatological impairments

PROGRESSIVE INCOORDINATION:

1. Brain tumors
2. metabolic conditions
3. ataxia
4. hydrocephalus
5. myoclonic epilepsy

EVALUATION FOR ASSOCIATED CONDITIONS

1. ADHDDCD is strongly associated with ADHD. Approximately 50 percent of kids with ADHD have DCD and vice versa. It is more associated with inattentive type of ADHD. and associated with fine motor skills deficit.
2. ASD...approximately 5% of kids with DCD have ASD. In ASD approximately 80% had definite and 10% have borderline motor impairment. The motor difficulties in children with ASD typically correlate with the degree of cognitive, social and emotional impairments and tend to be more severe than those in children with co-occurring DCD and ADHD.
3. Specific language impairment...in one observational study, approximately one third of children with SLI had DCD.
4. Learning disability...DCD co exists with non motor learning disabilities. Children with DCD may have difficulties in executive functioning and struggle with cognitive tasks such as nonverbal working memory, fluency, inhibition and planning. However children with DCD perform similarly to typically developing children in verbal tasks.



Why doesn't my child sit, doctor?

Dipanjana Datta^a PhD | Anjan Bhattacharya^b FRCPCH

Introduction

How many of us have heard this complaint from parents and our heart sank? If a child does not crawl in time or even, does not walk in time, we still think, they have a fighting chance. A child not even sitting behooves possible sinister prognosis.

Case 1

Anand was born after his parents had a deformed still born 18 months back! Six more months down the line, he was rushed in to your chamber with anxious parents with the complaint. You knew that developmental milestones can vary, so you adopted watchful expectancy! Anand started having recurrent coughs and colds and he stopped thriving! You ran basic tests, which were normal. Your felt reassured and treated his respiratory infections. When he was 10 months old, getting scrawny and floppier, you sought tertiary care help. Parents returned with a diagnosis but a dead second child!

Case 2

Shamim was still bottom shuffling when he was 1 year and you referred him to physiotherapist. When even after 2 more months, Shamim started deteriorating, you ran your thyroids and other tests, including MRI brain scan, which did not reveal much. You continued supportive care but Shamim continued to deteriorate. By around 2nd year, when Shamim became bed-bound, you pondered about consanguinity and asked a Genetic colleague, who ran some Genetic Tests. Shamim passed away from a fulminant influenza but parents could be counselled about their risk in their next pregnancy!

Case 3

Jyotsna is a girl, who started school with a mere limp. Parents could not conceive further and yet Jyotsna was not thriving soon after starting school. She started missing her schools from recurrent chest infections, became top heavy and was otherwise a very chirpy soul. With progression of age however, her performances started deteriorating and her respiratory infections started becoming too frequent! She could not join friends in play ground and gradually became wheelchair dependent over the years, despite of physiotherapy. One morning, parents found her in her room, passing away in her sleep following a relatively trivial "cold" and they were bewildered and distraught! Death in sleep is rather common in children, who harbours Spinal Muscular Dystrophy!!

The need to introduce screening for SMA in the new born screening program to facilitate early intervention and management

Spinal Muscular atrophy is a progressive neurodegenerative disease affecting the motor neurons in the anterior horn of the spinal cord. The progress of the disease is different from other neurodegenerative disorders as it has the greatest rate of motor strength and function loss at disease onset, followed by a slower rate in course, eventually, in many cases depending on the type, leading to early mortality. (7,8)

Depending on the age of onset and on the maximum age of motor function achieved in an individual, SMA can be classified into 5 types. The onset of symptoms for SMA type 0 is

-
- a. Consultant Genetic Counselor, Child Development Centre, Apollo Gleneagles Hospital, Kolkata & Organization of Rare Diseases, India
- b. Developmental Paediatrician, Child Development Centre, Apollo Gleneagles Hospital, Kolkata



prenatal. At birth the child is presented with joint contractures, respiratory distress, and diffused hypotonia. They do not survive the neonatal period without intervention.

Infants with SMA type 1 (SMA1) is presented at birth to 6 months of life with hypotonia, difficulty breathing or feeding, and motor delays. In addition to life-threatening pulmonary complications, individuals with SMA1 often have bulbar dysfunction and dysphagia because of muscle weakness. As a result, they may have nutritional compromise and failure to thrive. These patients never sit and have a life expectancy of < 2 years without intervention. The primary cause of death is pulmonary compromise, because of respiratory muscle weakness leading to severe restrictive lung disease and progressive respiratory failure.

Infants with SMA type 2 (SMA2), usually have a disease onset around 6 months but before age 18 months. These infants are able to sit but do not reach the motor milestone of walking and survive into early adulthood.

Children with SMA type 3 (SMA3) have onset around 18 months of age and achieve to stand independently and may also be able to walk. They have regression in their milestones and in teens may lose the ability to walk. They survive well into adulthood.

Finally, individuals with SMA type 4 (SMA4) present at adulthood and progress very slowly. These patients stand, may walk, and usually have normal lifespan.(9,6)

The disease is caused by abnormalities in the SMN1 gene on chromosome 5q and is inherited in an autosomal recessive manner in most cases. The SMN1 gene product is crucial for motor neuron development. SMN is a ubiquitously expressed protein whose expression is reduced in SMA. SMN is required for the assembly of the small nuclear ribonucleoprotein (snRNP) complexes that mediate splicing (12; 13). snRNP assembly is defective in SMN-deficient SMA cells. snRNP assembly is more markedly reduced in

SMA mouse neural tissues than in other tissues like the kidney (14) suggesting that motor neurons are more sensitive to deficits in snRNP assembly. SMN may also have a function that is unique to motor neurons.

In approximately 95% of patients, SMA results from homozygous deletion or conversion of SMN1. In about 2% of patients, de novo deletions occur in one of the SMN1 alleles; in 3%-4%, other mutations can be found, typically with an SMN1 exon 7 deletions on the other allele. The point mutation found in homozygous or compound heterozygous states other than exon 7 in SMN1 has also been reported in 0.05% of population. These exons include exon 3, exon 6, exon 4 etc.. (10, 11)

In humans a large tandem chromosomal duplication has led to a second copy of the SMN gene locus known as SMN2. SMN2 is different from SMN1 by a single nucleotide that disrupts a splice enhancer in exon 7. Most of SMN2 mRNAs have a 5 bp difference in exon7 of SMN1 (SMN17) and this leads to production of an unstable and non functional/ less than functional protein. Though, this leads to 10-20% of the functional SMN2 gene product, it is well known that increased genomic copies of SMN2 inversely correlates with disease severity among individuals with SMA. Genetic diagnostics would hence involve MLPA for SMN 1/ SMN2 gene and if this MLPA is non-informative, gene sequencing for SMN 1 is needed. (10,11)

Presence of additional modifiers, other than SMN2 that can influence the phenotype positively as well as negatively. These lead to a spectrum of phenotype within the subtypes also. (11)

Understanding of the disease and its pathway has helped in emergence of new therapies. Nusinersen, commercially known as Spinraza is an antisense oligonucleotide that is directly administered into the cerebrospinal fluid. In 2016, it became the first drug to be approved for treatment of SMA in paediatric and adult patients.



Spinraza works by blocking the splicing of SMN2 pre-mRNA; this results in the inclusion of exon 7 into the final protein. Since, SMN1 and SMN2 vary by a single exon, i.e., exon 7, this modification of the SMN2 pre-mRNA results in the formation of a functional SMN protein that can reduce the severity of SMA. Cherish, a phase 3 Spinraza trial, was conducted with 126 children (84- nusinersen, 42- control) aged 2 to 9 years with late-onset SMA (Types 2 and 3). It was a 15-month long, randomized sham-controlled study that analyzed the effects of Spinraza on motor function.

The experimental group saw a significant improvement in the Hammersmith Functional Motor Scale—expanded, which was the primary endpoint, and Revised Upper Limb Module scores(17).

Another drug in the horizon is, Onasemnogene abeparvovec-xioi, or more commonly known by its trade name Zolgensma, is a one-time adeno-associated viral vector-based gene therapy used for treating SMA Type 1 in children below the age of 2 years.

Administered intravenously, this therapy works by introducing a functional copy of the SMN gene in the motor neurons. The efficacy of this gene therapy was studied in two open-label clinical trials. A 24-month trial START evaluated the effectiveness and safety of the drug in 15 patients, who were divided into high- and low-dose cohorts with 12 and 3 patients each, respectively. At the end of the 24-month trial period, 11/12 patients from the high-dose cohort were able to independent sitting while all of them were able to avoid ventilation. Achievement of independent sitting and event-free survival were the primary endpoints for the STRIVE trial that enrolled 22 patients. Approximately 47.6% of patients were able to sit independently and 91% of patients did not require breathing support. (15)

Also recently approved by FDA is the drug, Risdiplam, marketed as Evrysdi is an oral therapy for SMA that was approved by the FDA in August 2020. It can be administered in patients

aged 2 months and older. It is a SMN2 pre-mRNA splicing modifier that is taken as liquid. By avoiding interaction with the human multidrug resistance protein 1, it distributes to the central nervous system and peripheral tissues. The efficacy of this therapy was evaluated in two trials: FIREFISH and SUNFISH. FIREFISH was an open-label pivotal study in 21 infants aged 1 to 7 months with Type 1 SMA. Forty one percent of the infants were able to meet the primary goal of the trial, which was being able to sit without support for five seconds. A four-point or more increase in CHOP-INTEND scale was also found in approximately 90% of the patients. SUNFISH is a multicentre, placebo-controlled study that aims to assess the effectiveness and safety of Evrysdi in SMA type 2 and 3 patients aged 2 to 25 years. The study is divided into 2 parts, with 51 patients enrolled in part 1 and 180 non-ambulatory patients in part 2. The primary endpoint of part 2 of the study is a significant change in the Motor Function Measure-32 (MFM32) scale score after 12 months of drug usage. Data from the second part showed that children aged 2 to 5 years, responded better to the treatment and 78.1% of children treated with Evrysdi showed a three-point or more improvement in the MFM32 scale score.(16)

Because of the above criteria where we have a good knowledge of the disease, and a well established genetic diagnostic algorithm along with emerging new therapies, new born screening of infants for SMA is a viable option. The expense of the diagnostic test and therapy though high, however, early detection paves the way to early intervention.

In the present moment the management of SMA is done through international standard of care guidelines formed by consensus statement of 60 experts working with this disorder.(3) The management of SMA, until recently had consisted primarily of supportive care to slow or prevent respiratory failure, to address the nutritional issues, to monitor or slow on set of scoliosis and joint contractures. Respiratory care involves use of devices that give ventilation support, especially during sleep and viral illnesses when hypoventilation is most likely to occur, as well as methods to mechanically augment cough



and clearance of respiratory secretions.(1) Nutritional support includes the use of non oral methods to deliver enteral nutrition, typically through a surgically placed feeding tube or temporary nasal tube, plus medical or surgical interventions to control gastroesophageal reflux. (1,2,3,) Management of joint contractures and scoliosis involves aggressive physical therapy assessments (CHOP-Intend, HMSME, HINES, Bayleys, RULM) , daily passive range of motion exercises and use of bracing to facilitate and maintain optimal positioning of extremities and maintain the spine upright against gravity. Surgical intervention with internal fixation of the spine may also be needed.(1, 2,3,4,5) Monitoring the cardiac activities and also issues like GERD are also addressed.

Although the incidence of SMA internationally is reported at 1 in 11,000 births (8), the actual epidemiology numbers in India are not known. It is nevertheless the most common fatal genetic disease of infancy.

New born screening accelerates early diagnosis. Early diagnosis facilitates early intervention through management even before the onset of symptoms. This leads to a better quality of life.

It should be noted that while, SMA type 0, 1 and 2 are severe and life threatening, SMA type 3 and Type 4 are milder forms. Individuals with SMA type 3 and type 4, often lead a family life. Early supportive intervention can help to prolong life and slow the natural history of SMA in increased lifespan and help in improving the quality of life. This also gives the families a chance to seek time for the therapies that are evolving.

Key Learning Points

1. Anand, Shamim, Jyothnas matter – their Quality of Life (QoL) matter!
2. We, as Paediatricians, can save most of these children through a. Newborn Screening, b. Early Detection, c. Early (expert) Intervention and d. (Expert) Genetic Counseling
3. Relatively inexpensive, high quality Genetic Tests are available in all metropolises and many second tier cities in India by now
4. A Genetic i.e. accurate diagnosis can lead to medication and appropriate multidisciplinary management, prolonging and maintaining good QoL

5. Expert Genetic Counseling can save and secure the next child/progeny

Reference

1. Kolb SJ, Coffey CS, Yankey JW, et al; NeuroNEXT Clinical Trial Network on behalf of the NN101 SMA Biomarker Investigators. Natural history of infantile-onset spinal muscular atrophy. *Ann Neurol*. 2017;82(6):883-91.
2. Iannaccone ST. Modern management of spinal muscular atrophy. *J Child Neurol*. 2007;22:974-78.
3. Mercuri E, Finkel RS, Muntoni F, et al; SMA Care Group. Diagnosis and management of spinal muscular atrophy: part 1: recommendations for diagnosis, rehabilitation, orthopedic and nutritional care. *Neuromuscul Disord*.2018;28(2):103-15.
4. DiVito D, Konek S. Spinal muscular atrophy: summary for nutritional care. *Infant Child Adoles Nutr*. 2010;2(6):348-54.
5. Wang CH, Finkel RS, Bertini ES, et al; Participants of the International Conference on SMA Standard of Care. Consensus statement for standard of care in spinal muscular atrophy. *J Child Neurol*. 2007;22(8):1027-49.
6. Vamshi K. Rao, MBBS, MD; Daniel Kapp, PharmD, BCPS; Mary Schroth, MD; Gene Therapy for Spinal Muscular Atrophy: An Emerging Treatment Option for a Devastating Disease, *JMCP* , Supplement to *Journal of Managed Care & Specialty Pharmacy* Vol. 24, No. 12-a December 2018
7. V Cusin, O Clermont, B Gérard, D Chanterreau, J Elion, *Med Genet* 2003;40:e39
8. Pearn J. Incidence, prevalence, and gene frequency studies of chronic childhood spinal muscular atrophy. *J Med Genet* 1978;15:409-13.
9. Campbell L, Potter A, Ignatius J, Dubowitz V, Davies K. Genomic variation and gene conversion in spinal muscular atrophy: implications for disease process and clinical phenotype. *Am J Hum Genet* 1997;61:40-50.
10. Chen KL, Wang YL, Rennert H, Joshi I, Mills JK, Leonard DG, Wilson RB. Duplications and de novo deletions of the SMNt gene demonstrated by fluorescence-based carrier testing for spinal muscular atrophy. *Am J Med Genet* 1999;85:463-9
11. Clermont O, Bürglen L, Burlet P, Lefebvre S, Viollet L, Hausmanowa-Petrusewicz I, Munnich A Melki J. Genetics analysis of unusual SMA pedigrees using SMN, NAIP genes and markers C212-C272. *Am J Hum Genet* 1995;57:A237.
12. Pellizzoni,L.(2007).Chaperoningribonucleo protein biogenesisin health and disease. *EMBORep*. 8,340-345.doi:10.1038/sj.embor. 7400941
13. Burghes,A.H.M.,andBeattie,C.E.(2009).



Journal Scan

Human milk oligosaccharide 2'-fucosyllactose links feedings at 1 month to cognitive development at 24 months in infants of normal and overweight mothers

Paige K. Berger et al.

PLOS ONE | <https://doi.org/10.1371/journal.pone.0228323> 12 Feb 20

Abstract

Background : Infant cognitive development is influenced by maternal factors that range from obesity to early feeding and breast milk composition. Animal studies suggest a role for human milk oligosaccharide (HMO), 2'-fucosyllactose (2'FL), on learning and memory, yet no human studies have examined its impact on infant cognitive development relative to other HMOs and maternal factors.

Objective : To determine the impact of 2'FL from breast milk feeding on infant cognitive development at 24 months of age relative to maternal obesity and breast milk feeding frequency.

Methods and materials : Hispanic mother-infant pairs (N = 50) were recruited across the spectrum of pre-pregnancy BMI. Breast milk was collected at 1 and 6 months, and feedings/day were reported. Nineteen HMOs were analyzed using high-performance liquid chromatography, with initial interest in 2'FL. Infant cognitive development score was assessed with the Bayley-III Scale at 24 months. Linear regressions were used for prediction, and bootstrapping to determine mediation by 2'FL.

Results : Maternal pre-pregnancy BMI was not related to feedings/day or HMOs, but predicted poorer infant cognitive development ($\beta = -0.31$, $P = 0.03$). Feedings/day ($\beta = 0.34$) and 2'FL ($\beta = 0.59$) at 1 month predicted better infant cognitive development (both $P < 0.01$). The association of feedings/day with infant cognitive development was no longer significant after further adjustment for 2'FL (estimated mediation effect = 0.13, $P = 0.04$). There were no associations of feedings/day and 2'FL at 6 months with infant cognitive development.

Conclusions : Our findings suggest that maternal factors influence infant cognitive development through multiple means. Though maternal obesity may be a separate negative influence, greater frequency of breast milk feeding at 1 month contributed to infant cognitive development through greater exposure to 2'FL relative to other HMOs. The influence of 2'FL was not significant at 6 months, indicating that early exposure to 2'FL may be a critical temporal window for positively influencing infant cognitive development.



Breastfeeding Duration Is Associated with Regional, but Not Global, Differences in White Matter Tracts

Christopher E. Bauer et al.

Brain Sci. 2020, 10, 19; doi:10.3390/brainsci10010019

Abstract

Extended breastfeeding through infancy confers benefits on neurocognitive performance and intelligence tests, though few have examined the biological basis of these effects. To investigate correlations with breastfeeding, we examined the major white matter tracts in 4–8 year-old children using diffusion tensor imaging and volumetric measurements of the corpus callosum. We found a significant correlation between the duration of infant breastfeeding and fractional anisotropy scores in left-lateralized white matter tracts, including the left superior longitudinal fasciculus and left angular bundle, which is indicative of greater intrahemispheric connectivity. However, in contrast to expectations from earlier studies, no correlations were observed with corpus callosum size, and thus no correlations were observed when using such measures of global interhemispheric white matter connectivity development. These findings suggest a complex but significant positive association between breastfeeding duration and white matter connectivity, including in pathways known to be functionally relevant for reading and language development.



Journal Scan

Factors associated with exclusive breastfeeding at hospital discharge: a study using data from the Georgian Birth Registry

Marie Sigstad Lande et al.

International Breastfeeding Journal (2020) 15:39. (<https://doi.org/10.1186/s13006-020-00286-9>)

Abstract

Background: The World Health Organization recommends exclusive breastfeeding for six months, defined as no other solids or liquids besides breast milk and essential vitamins or medicines. Data about exclusive breastfeeding are limited in Georgia, and the information that exist are provided by national surveys, that present inconsistent numbers. Georgia has recently established a national birth registry, which includes information about early postpartum breastfeeding. The objective of this study was to identify factors associated with exclusive breastfeeding of term newborns at hospital discharge in Georgia, using national registry data.

Methods: All live, singleton, term births registered in the Georgian Birth Registry in November and December 2017 were included, with a final study sample of 7134 newborns. Newborns exclusively breastfed at hospital discharge were compared with those who were not, and potential factors were assessed with logistic regression analysis. Hospital discharge normally occurred between 2 and 5 days postpartum.

Results: The study identified several factors associated with nonexclusive breastfeeding of term newborns at hospital discharge in Georgia: maternal higher education compared to secondary education or less (Adjusted Odds Ratio [AOR] 0.75; 95% CI 0.59, 0.97), caesarean delivery compared to vaginal or assisted vaginal delivery (AOR 0.47; 95% CI 0.37, 0.60), birthweight < 2500 g compared to 3000–3499 g (AOR 0.51; 95% CI 0.27, 0.97), and admission to neonatal intensive care unit after delivery (AOR 0.02; 95% CI 0.02, 0.03). None of the following factors were associated with exclusive breastfeeding at discharge: mother's age, marital status, Body Mass Index (BMI), parity, in vitro fertilization, maternal intrapartum complications and the sex of the newborn.

Conclusions: To the authors' knowledge, this is the first time determinants of exclusive breastfeeding at hospital discharge have been studied in Georgia. Several factors associated with nonexclusive breastfeeding at discharge were identified, most noteworthy were caesarean delivery and admission to neonatal intensive care unit. These findings are of importance to the Georgian health authorities and maternal/child non-governmental organizations.



Quiz

Dr. Arun Prasad

Joint Secretary, IAP NDD Chapter

Secretary, IAP Chandigarh

Director, Asha Child Care And Development Centre

MILESTONES

1. WHICH OF THE FOLLOWING IS THE EXAMPLE OF “DOUBLE SYLLABLE BABBLE” DISPLAYED BY AN INFANT AROUND 9-12 MONTHS

- a) Ma-da
- b) Ma-ba
- c) Ba-da
- d) Ba-ba
- e) Ba-ma

2. IN THE NORMAL FINE MOTOR DEVELOPMENT WHICH OF FOLLOWING SHOULD OCCUR FIRST ?

- a) Copying a circle
- b) Copying a cross
- c) Drawing a square
- d) Tripod pencil grip
- e) Copying a triangle

3. AT WHAT AGE SHOULD A CHILD DEVELOP A MATURE PENCIL GRIP (Dynamic Tripod Grasp) ?

- a) 2 years
- b) 3 years
- c) 4 years
- d) 5 years
- e) 6 years

4. AROUND WHAT AGE A CHILD SHOULD BE ABLE TO BUILD A TOWER OF THREE BUILDING BLOCKS?

- a) 9 months
- b) 12 months
- c) 15 months
- d) 18 months
- e) 21 months

PSYCHOLOGICAL

5. WHICH SPECIFIER IS LEAST LIKELY IN CHILDHOOD SPECIFIC PHOBIAS ?

- a) Animal phobias
- b) Natural environment phobias
- c) Blood/injection/injury phobias
- d) Situational phobias

6. DIAGNOSTIC CRITERIA FOR PANIC DISORDER INCLUDES ALL OF THE FOLLOWING EXCEPT:

- a) An abrupt surge of intense fear or intense discomfort that reaches a peak within minutes
- b) Expected Panic Attacks
- c) Persistent concern about panic attacks
- d) Significant maladaptive change in behavior related to attacks

7. WHAT IS NOT A RISK FACTOR FOR SELECTIVE MUTISM?

- a) Overprotective and controlling parents
- b) History of shyness
- c) Parent with Social Anxiety
- d) Parent with Major Depression



Quiz

8. A MAJOR ETIOLOGICAL FACTOR FOR SEPERATION ANXIETY IS :

- a) Peer bullying
- b) Inconsistent and ? or overtly intrusive parenting
- c) Traumatic experience of a peer
- d) Major diagnosis of bipolar disorder

ADHD

9. NEUROTRANSMITTERS IMPLICATED IN CAUSE OF ADHD INCLUDE ALL OF THE FOLLOWING EXCEPT :

- a) Dopamine
- b) Glutamate
- c) Epipherine
- d) GABA

10. RECENT RESEARCH HAS SHOWN THAT

- a) ADHD is found equally among boys and girls
- b) ADHD is caused by single gene called DAT 1

- c) The brains of children with ADHD are 3-4% smaller than children without ADHD
- d) The same medicines used to treat depression are the most effective to treat ADHD

11. WHAT ARE THE THREE TYPES OF SYMPTOMS FOR ADHD ?

- a) Hyperactivity , impulsivity, inattention
- b) Inattention, distraction, hyperactivity
- c) Inattention , impulsivity, distraction
- d) Hypervigilance, increased startle and distraction

12. WHICH OF FOLLOWING IS NOT A CHARECTERISTIC OF ADHD ?

- a) Inattention
- b) Learning disability
- c) Self control
- d) Fidgety

Please send answers to lata2207@gmail.com / Kawaljit000@gmail.com before 25 september 2020. Correct answer will be published in next issue

Answers - JULY

- | | |
|------|-------|
| 1. C | 6. C |
| 2. A | 7. A |
| 3. D | 8. C |
| 4. A | 9. D |
| 5. B | 10. D |

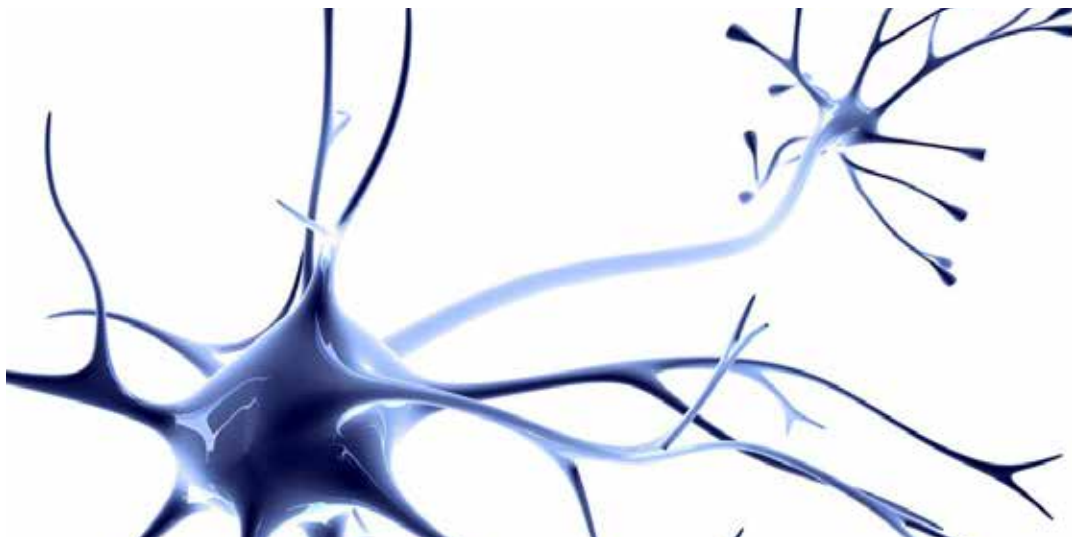


Month in pics

ACADEMY OF PEDIATRIC NEUROLOGY CHAPTER



NEURO-NEURODEVELOPMENT CHAPTER WEBINAR SESSION 19



DEAR MEMBERS,
GREETINGS FROM IAP!


Join us for a very enriching discussion on **neurological problems associated with neurodevelopmental disorders**

This webcast is brought to you under dIAP, an initiative of IAP to facilitate e-learning in all spheres of pediatrics. Live webcast of the webinar discussions, on-line clinics and their subsequent archiving is one of the activities under this banner.

MODERATORS

- | | | | |
|---|--|--|---|
| 
Dr Vasant Khalatkar | 
Dr Lokesh Lingappa | 
Dr Chetan Shah | 
Dr Kawaljit Multani |
|---|--|--|---|

EXPERTS

- | | | |
|--|--|---|
| 
Dr MKC Nair | 
Dr Sameer Dalwai | 
Dr Velmurugan |
| 
Dr Chandrika Rao | 
Dr Vrishab Gawli | |

Go to diapindia.org/event-calendar or [click here](#)

With warm regards
DR BAKUL JAYANT PAREKH
DR GV BASAVARAJ

DATE | WEDNESDAY, SEP 2
TIME | 8 PM TO 10 PM

If you are not able to view on the above link, [please click here](#)



Month in pics



Meeting ID : 818 6134 1880

Password : Cognicare



LIVE

Webex Meetings

In Association with IAP South Delhi

My 2 year old has not started to talk – Approach to case of Language Delay in office Practice

August'20

20

Thursday

TIME :

9.00 PM - 10.30 PM

Moderator



Dr Puja Grover Kapoor

Consultant Pediatric Neurologist,
Rainbow Children Hospital, Delhi,
Paras Hospital, Gurgaon.

Speaker



Dr Shambhavi Seth

Sr. Consultant Developmental
Pediatrician
Max Hospital, Saket & Gurgaon
BLK Super Speciality Hospital,
Bright Beginning CDC, New Delhi.

Secretary of IAP South Delhi



Dr Manu Agarwal

Sr. Consultant Paediatrician,
Paediatric Endocrinologist at
Max & Sitaram Bhartia Hospitals.

President of IAP South Delhi



Dr Praveen Khilnani

MBBS, MD, FAAP, MCCM (USA),
Diplomat American Board of Pediatrics
& Critical Care Medicine. Director PICU
& Group Head-Academics Sr. Consultant
Pediatrics & Pediatric Pulmonology

Please Click to join the Webinar

Host by
Brio Bliss Life Science Pvt. Ltd.
Makers of
CogniCare™

For any query please whatsapp 99712 94128





Many Teachers

Sanskrit words for 'teacher' based on their unique abilities...

- 1. The teacher who gives you information is called: *Adhyapak.*
2. The one who imparts knowledge combined with information is called: *Upadhyaya.*
3. The one who imparts skills is called: *Acharya.*
4. The one who is able to give a deep insight into a subject is called: *Pandit.*
5. The one who has a visionary view on a subject and teaches you to think in that manner is called: *Dhrishita.*
6. The one who is able to awaken wisdom in you, leading you from darkness to light, is called: *Guru.*

Sanskrit is, perhaps, the only language that has such a refined vocabulary to distinguish the different kinds of teachers.